

UNMATCHED LEFT PARENTHESIS '(SAPOSIN'

The number of right parentheses in a query must be equal to the number of left parentheses.

=> s saposin (p) blood

L9 44 SAPOSIN (P) BLOOD

=> d ibib abs 1-44

L9 ANSWER 1 OF 44 CAPLUS COPYRIGHT 2004 ACS on STN

ACCESSION NUMBER: 2004:941423 CAPLUS

TITLE: Immunoquantification of α -galactosidase:

Evaluation for the diagnosis of fabry disease

AUTHOR(S): Fuller, Maria; Lovejoy, Melanie; Brooks, Doug A.;

Harkin, Miriam L.; Hopwood, John J.; Meikle, Peter J.

CORPORATE SOURCE: Lysosomal Diseases Research Unit, Department of Genetic Medicine, Women's and Children's Hospital, North Adelaide, Australia

SOURCE: Clinical Chemistry (Washington, DC, United States) (2004), 50(11), 1979-1985

CODEN: CLCHAU; ISSN: 0009-9147

PUBLISHER: American Association for Clinical Chemistry

DOCUMENT TYPE: Journal

LANGUAGE: English

AB Background: Fabry disease is an X-linked inborn error of glycosphingolipid

catabolism resulting from a deficiency of the lysosomal exoglycohydrolase,

α -galactosidase. Enzyme replacement therapy is currently available for Fabry disease, but early diagnosis before the onset of irreversible pathol. will be mandatory for successful treatment. Presymptomatic detection would be possible through the use of a newborn-screening program. We report on the use of sensitive assays for the measurement of α -galactosidase protein and activity and for the protein **saposin C**, which are diagnostic markers for Fabry disease.

Methods: Two sensitive immunoassays for the measurement of α -galactosidase activity and protein were used to determine the concns. of α -galactosidase in dried filter-paper **blood spots** and plasma samples from control patients and patients with a lysosomal

storage

disorder (LSD). Results: Fabry hemizygous individuals were clearly identified from control populations by decreases in both α -galactosidase activity and protein. Fabry heterozygotes generally fell between the hemizygotes and controls. Including the measurement of **saposin C** enabled differentiation between Fabry heterozygotes and controls. In **blood spots**, all Fabry individuals could be distinguished from control **blood spots** as well as from 16 other LSD patients. Conclusions: The determination of α -galactosidase

activity or

protein in dried filter-paper **blood spots** could be used for the diagnosis of Fabry patients. With further validation, these assays could be used for the identification of Fabry patients in newborn-screening programs and may also be suitable for screening high-risk populations.

REFERENCE COUNT: 19 THERE ARE 19 CITED REFERENCES AVAILABLE FOR THIS

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ACCESSION NUMBER: 2004:817730 CAPLUS

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=> s saposin

L8 1288 SAPOSIN

=> s (saposin (p) (blood or serum or urine or amniotic)

UNMATCHED LEFT PARENTHESIS '(SAPOSIN'

The number of right parentheses in a query must be equal to the
number of left parentheses.

=> s (saposin (p) (blood)

3 FILES SEARCHED...

L10 646 SAPOSIN (1W) (A OR C OR D)

=> s ((saposin) (1w) (a or c or d)) (p) blood

3 FILES SEARCHED...

L11 28 ((SAPOSIN) (1W) (A OR C OR D)) (P) BLOOD

=> d ibib abs 1-28

L11 ANSWER 1 OF 28 CAPLUS COPYRIGHT 2004 ACS on STN

ACCESSION NUMBER: 2004:941423 CAPLUS

TITLE: Immunoquantification of α -galactosidase:

Evaluation for the diagnosis of fabry disease

AUTHOR(S): Fuller, Maria; Lovejoy, Melanie; Brooks, Doug A.;

Harkin, Miriam L.; Hopwood, John J.; Meikle, Peter J.

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SOURCE: Clinical Chemistry (Washington, DC, United States)
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α -galactosidase. Enzyme replacement therapy is currently available
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detection would be possible through the use of a newborn-screening
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blood spots could be used for the diagnosis of Fabry patients.

With further validation, these assays could be used for the

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of Fabry patients in newborn-screening programs and may also be suitable
for screening high-risk populations.

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L11 ANSWER 2 OF 28 CAPLUS COPYRIGHT 2004 ACS on STN

that this precursor cell in the digesting macrophage system also has an impaired metabolic catabolism for lipopigments (3). Immunohistochemical studies indicate that microglial reaction in NCL brain is limited to resident microglia without contribution by circulating monocytes (4). The granular osmiophilic deposit (GROD) type of NCL has now been established not only in infantile, but also in late-infantile, juvenile, and protracted-juvenile NCL (5). A European Tissue Registry established within the framework of a European Concerted Action on Neuronal Ceroid-Lipofuscinosis may form the basis for additional collaborative studies on NCL, including both biopsy and autopsy tissues.

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ACCESSION NUMBER: 90030178 EMBASE
DOCUMENT NUMBER: 1990030178
TITLE: Sphingolipid hydrolase activator proteins and their precursors.
AUTHOR: Sano A.; Hineno T.; Mizuno T.; Kondoh K.; Ueno S.; Kakimoto Y.; Inui K.
CORPORATE SOURCE: Department of Neuropsychiatry, Ehime University School of Medicine, Ehime 791-02, Japan
SOURCE: Biochemical and Biophysical Research Communications, (1989) 165/3 (1191-1197).
ISSN: 0006-291X CODEN: BBRCA
COUNTRY: United States
DOCUMENT TYPE: Journal; Article
FILE SEGMENT: 025 Hematology
029 Clinical Biochemistry
LANGUAGE: English
SUMMARY LANGUAGE: English

AB Activator proteins for sphingolipid hydrolases (**saposins**) are small acidic, heat-stable glycoproteins that stimulate the hydrolysis of sphingolipids by lysosomal enzymes. The molecular mass of each stimulator is about 10 kDa, but glycosylated forms of higher mass exist too. The distribution and developmental changes in two **saposins** and their precursor proteins were studied with the aid of monospecific antibodies against **saposin-B** and **saposin-C**. They show a wide distribution in rat organs and forms intermediate between **saposin** and prosaposin (the precursor protein containing four different **saposin** units) could be seen. The amount of **saposin** and the degree of processing from prosaposin are quite different in different tissues. The **saposins** are the dominant forms in spleen, lung, liver, and kidney, while skeletal muscle, heart, and brain contain mainly precursor forms. In human **blood**, leukocytes contain mainly **saposin**, while plasma contains mainly precursor forms and platelets show many forms. Their subcellular distribution was studied using rat liver. The **saposins** of approximately 20 kDa are dominant in the light mitochondrial, mitochondrial, and microsomal fractions, following the distribution of the activity of a lysosomal marker enzyme. The nuclear fraction exhibits bands corresponding to non-glycosylated **saposin**. The soluble fraction contained much precursor forms. A developmental study of rat brain showed that the concentration of **saposin** precursors increased with age.

=> saposin (1w) (a or c or d)

developed by creating a null allele in embryonic stem cells through gene targeting to investigate the phenotypic diversity of prosaposin mutations and the involvement of this protein in lysosomal storage diseases, and for the development of therapeutic approaches. Mice homozygous mutants die at the age of 35-40 days and neurological disorders contribute to the early demise of the mutant mice. The male reproductive organs in homozygous mutants show several abnormalities, such as a decrease in testis size with reduced spermiogenesis and an involution of the prostate, seminal vesicles, and epididymis. In these animals, the **blood** levels of testosterone remain normal. In the prostate of homozygous mutants, only the basal epithelial cells appear to be present, while the secretory cells are absent. These findings suggest that prosaposin may be involved in the development and maintenance of the male reproductive organs, as well as, in cellular differentiation.

L11 ANSWER 22 OF 28 MEDLINE on STN
 ACCESSION NUMBER: 90121224 MEDLINE
 DOCUMENT NUMBER: PubMed ID: 2610686
 TITLE: Sphingolipid hydrolase activator proteins and their precursors.
 AUTHOR: Sano A; Hineno T; Mizuno T; Kondoh K; Ueno S; Kakimoto Y; Inui K
 CORPORATE SOURCE: Department of Neuropsychiatry, Ehime University School of Medicine, Japan.
 SOURCE: Biochemical and biophysical research communications, (1989 Dec 29) 165 (3) 1191-7.
 Journal code: 0372516. ISSN: 0006-291X.
 PUB. COUNTRY: United States
 DOCUMENT TYPE: Journal; Article; (JOURNAL ARTICLE)
 LANGUAGE: English
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 ENTRY MONTH: 199002
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on STN

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TITLE: Immunoquantification of α -galactosidase: Evaluation for the diagnosis of fabry disease.
AUTHOR: Fuller M.; Lovejoy M.; Brooks D.A.; Harkin M.L.; Hopwood J.J.; Meikle P.J.
CORPORATE SOURCE: M. Fuller, Lysosomal Diseases Research Unit, Department of Genetic Medicine, Women's and Children's Hospital, 72 King William Rd., North Adelaide, SA 5006, Australia. maria.fuller@adelaide.edu.au
SOURCE: Clinical Chemistry, (2004) 50/11 (1979-1985).
Refs: 19
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